CASE REPORT

A Rare Case of a Recurrent Giant Solitary Fibrous Tumor of the Ciliary Body of the Orbit

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Abstract Solitary fibrous tumors (SFTs) are uncommon spindle-cell tumors of mesenchymal origin initially described in the pleura and subsequently in other extra-pleural sites. These tumors are categorized as tumors of ‘intermediate malignancy’ under the World Health Organization classification of soft tissue tumors. SFT was virtually non-existent or misdiagnosed until its characteristic features, particularly the strong and diffuse immuno-reactivity to CD 34 were described. Extra-pleural manifestations of SFT, particularly in the head and neck region are rare. Although a number of isolated case reports of orbital SFTs have been described ever since its initial description in 1994, cases of recurrent SFTs have been very few. Recurrences of these tumors following surgery are considered unusual and metastasis exceptional. We describe clinical presentation and the management challenges of recurrent giant orbital SFT in a 15-year old girl along with a brief review of literature

Keywords Orbital tumors · Ciliary body tumor · Solitary fibrous tumor · Local recurrence

Introduction

Solitary fibrous tumors (SFTs) are uncommon spindle-cell tumors of mesenchymal origin [1]. They are most commonly reported to arise from the pleura and also from various other extra-pleural sites, including the

mediastinum, lung, liver, breast, retroperitoneum, spine, meninges and extra-cranial head and neck regions such as the orbit, sino-nasal cavity, salivary gland, thyroid gland and along the upper aero-digestive tract [2]. We describe the clinical presentation and further discuss the management challenges of a rare case of recurrent giant orbital SFT in a 15-year old girl along with a brief review of literature.

Case Report

A 15-year-old girl presented to our centre with a recurrent swelling of her left orbit for 2 years. She gave a history of a surgical debulking for a similar tumour of her left orbit 3 years prior at another centre. An increase in the residual tumour was appreciated by the patient a year following her initial surgery. A progressive increase in tumour size with associated pain and worsening vision prompted her to seek medical attention at our center.

Clinical examination revealed a large firm, non-pulsatile, non-compressible and mildly tender extra-ocular fleshy mass in the upper half of the left orbital cavity which resulted in proptosis and downward displacement of the left eye (Fig. 1). The skin over the left eyelid was stretched, edematous with prominent dilated veins. There were no ocular movements and the visual acuity in the left eye was restricted to perception of light. General physical examination, examination of the right eye and the rest of the head and neck was within normal limits. An MRI of the orbit showed an ill-defined, heterogenous mixed intensity lesion in the upper part of the left orbit involving the extraconal, conal and intraconal compartments with diffuse edema and dilated vessels in the left orbit. 28 mm of proptosis was noted. The fat planes

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Fig. 1 Clinical photograph at presentation

between the mass and the superior rectus, superior oblique and the anterior parts of the lateral and medial recti were indistinct. The mass caused mild compression along the postero-superior surface of the left eye ball. An intermediate intensity lesion in the left eyeball extending along the optic disc was noted. The mass was seen abutting the supero-lateral surface of the retro-bulbar segment of the left optic nerve with loss of fat planes. Loss of fat planes was also noted between the mass and the left lacrimal gland. There was however no extension into the superior orbital fissure, optic nerve or any intracranial extension (Fig. 2a, b). A trucut biopsy of the lesion along with a review of the slides of the prior surgery was suggestive of a solitary fibrous tumor. The patient underwent an uneventful left orbital exenteration, which entailed removal of the entire orbit including the globe, upper eyelid and orbital contents. The area of the proposed exenteration incision was marked giving wide margins to

the tumour and sparing the uninvolved lower eyelid. The dissection was further carried down through the periorbita to expose the orbital rim all the way up to the orbital apex with the aid of a periosteal elevator. Care was taken to avoid inadvertent penetration of the lamina papyracea into the ethmoid sinus air cells. A curved haemostat was used to clamp the tissue around the orbital apex while the exenteration specimen was cut and delivered anterior to the clamp. A frozen section confirmation of the apical stump tissue was done to ensure tumor free margins. The orbit was lined with a split-thickness skin graft after securing haemostasis and ensuring no cerebrospinal fluid leak. The patient is being planned for formal orbital prosthetic rehabilitation (Fig. 3a–c). The final histopathology confirmed the diagnosis of SFT, arising from the ciliary body of the left orbit resected with clear margins (Fig. 4a–d).

Discussion

Solitary fibrous tumor (SFT) is a distinctive, although rare, pathologic entity first described as a case of primary spindle-cell tumor of the pleura by Klemperer and Rabin in 1931. These tumors are categorized as tumors of ‘intermediate malignancy’ under the World Health Organization (WHO) classification of soft tissue tumors [3]. Although once thought to be mesothelial in origin, SFT is now considered a tumor of mesenchymal fibroblast-like cell origin [1].

There has been an increasing incidence of the tumor in the extrapleural, extraserosal sites such as the lung, liver, breast, meninges and in the head and neck region [2]. The real incidence of SFTs of the orbit is not clear, but these tumors have been increasingly recognized ever

Fig. 2 a, b MRI of the orbit showed an ill-defined, heterogenous mixed intensity lesion in the upper part of the left orbit involving the extraconal, conal and intraconal compartments

Fig. 3 **a** Intra operative clinical photographs of orbital exenteration. **b** Tumor bed following exenteration. **c** Specimen photograph

Fig. 4 **a** Tumour arising from the pigmented outer layer of pars plana of the ciliary body. H&E $\times 40$ **b** Higher magnification of tumour arising from the pigmented outer layer of pars plana of the ciliary

body. H&E $\times 200$ **c** Sheets of tumour cells with hyalinized blood vessels in between. H&E $\times 200$. **d** Tumour cells showing positive reaction for CD 34. DAB $\times 200$

since its initial description in 1994. The various locations of orbital SFT include the intraconal and extraconal spaces of the orbit, [2] lacrimal gland, lacrimal sac and eyelid. The tumour in our patient was found to be arising from the pigmented outer layer of pars plana of the ciliary body.

Orbital SFTs are typically known to cause insidious, painless proptosis and mostly develop in patients around the fourth decade (range 9–76 years). These tumours predominantly localise in the superior part of the orbit and are usually not associated with features of pain or nerve entrapments [4]. Tumour invasion into adjacent bone or

soft tissue is also uncharacteristic although recurrent tumours have been reported to invade the surrounding tissues as was seen in our patient.

Orbital SFTs were virtually non-existent or misdiagnosed until its characteristic histological features, including the strong and diffuse immunoreactivity to CD 34 was described [1, 5]. SFTs can occasionally demonstrate great variability in its morphology and shares histologic features with other mesenchymal tumors, which may contribute to its misdiagnosis. In a case series of 41 cases of orbital tumors previously classified as hemangiopericytoma, fibrous histiocytoma, and giant cell angiofibroma, the authors had found all the tumors to have overlapping morphologic and immunohistochemical features, further suggesting that SFT be used as a possible encompassing terminology [6]. The differential diagnosis of SFTs based on histopathological features includes meningioma, fibrous histiocytoma, schwannoma and more importantly hemangiopericytoma. A differential diagnosis to hemangiopericytoma with SFT is critical because hemangiopericytomas have an aggressive clinical as compared to the indolent nature of SFTs. More recent molecular analytical studies have suggested that almost all SFTs harbour an NAB2-STAT6 fusion gene, which is considered specific to this tumour type [7].

Various imaging modalities such as ultrasonography, CT scan, magnetic resonance imaging, and magnetic resonance angiography have been described for SFT and can be useful adjuncts in diagnosis [8]. Although there are no specific diagnostic features, a solitary, ovoid and well-defined mass with strong enhancement after contrast injection is suggestive of this diagnosis.

The mainstay of management of SFTs (both primary and recurrent tumours) is surgery which entails a local excision without any adjuvant therapy [9]. Safe surgical excision without breaching of the tumour capsule is considered essential to reduce the risk of local recurrence [10]. There is no conclusive evidence supporting the role of adjuvant radiotherapy or chemotherapy.

Although local excision is usually curative, orbital SFT has occasionally been reported to recur and rarely metastasize [10]. Bernardini et al. [11] reported 8 recurrences in his review of 42 cases of orbital SFTs. Local recurrences are considered to be due to incomplete excision of the tumour and such tumours show a tendency to invade into the surrounding tissues and orbital bone, rendering complete re-excision more difficult [9].

In conclusion, SFTs should be considered as a differential when confronting a painless slowly growing orbital mass that induces globe displacement. A careful upfront surgical excision with clear margins is essential for adequate local control of the tumour.

Compliance with Ethical Standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical Issues All procedures performed in this case report were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Human and Animal Rights Statement “This article does not contain any studies with animals performed by any of the authors”.

Informed Consent “Informed consent was obtained from the individual participant included in this study” “Additional informed consent was obtained from the individual participant for whom identifying information is included in this article”.

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